INTRODUCTION

Lymphomas represent a group of malignant neoplasms of lympho-reticular origin which are divided into Hodgkin’s disease (HD) and non-Hodgkin’s lymphomas (NHL).1 According to REAL (revised European-American Lymphoma) classification, NHL is a heterogeneous group of diseases with peculiar, morphological, phenotypic and molecular features (B-cell neoplasms, T-cell and putative natural killer (NK)-cell neoplasms). The nasal cavities and paranasal sinuses are rarely affected by primary NHL. Common primary extra-nodal sites of lymphomas include liver, soft tissue, dura, bone, stomach, intestine, bone marrow and others.2

CASE REPORT

A 38 year old male reported to our department with the chief complaint of pain in the upper right posterior teeth since 5 months. Pain was sharp, continuous and radiated to the right temporal region. Patient had a habit of pan-masala (betel nut) chewing for the last 10-15 years and had quit the habit since one month. Upon general physical examination patient gave the history of loss of vision (right eye) since 4 months and unilateral exophthalmos involving the right eye (Fig 1). Patient had nasal obstruction (right side) since 1.5 years. Extra oral examination revealed swelling antero- posteriorly from ala of the nose up to 4cm before the tragus of the ear, superio-inferiorly from infra-orbital margin up to 3cm below the ala-tragal line. The swelling was tender on palpation, soft to firm in consistency and temperature of the overlying skin was elevated. Right submandibular lymph nodes were palpable, movable and tender. Intraoral examination revealed buccal vestibular obliteration i.r.t. 15 and well circumscribed swelling present palatally i.r.t. 15 extending upto 18, approx.1x1 cm and was tender (Fig 2). On the basis of history and clinical examination a provisional diagnosis of residual cyst with differential diagnosis of deep fungal infections and benign tumors were considered.

Intra-oral periapical radiograph, maxillary occlusal radiograph and OPG revealed radiolucent area periapical to 15, 16, 17 region. Fine needle aspiration of fluid revealed presence of lymphocytes in clumps, numerous spindle shaped cells dispersed throughout, abnormal mitotic figures, abnormal fragmentation of the nucleus and abnormal epithelial cells noted in the smear. Radiograph PNS (water’s view) shows large opacity occupying the right maxillary sinus extending

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MAXILLARY SINUS LYMPHOMA WITH VISION LOSS AND INTRAORAL PRESENTATIONS: A CASE REPORT

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medially into the oropharynx and superiorly into the base of the right orbit (Fig 3). Radiograph chest (PA view) was normal thus ruled out metastasis. CT had revealed a large soft tissue density mass measuring approx 7x6 cms in size, seen in the right maxillary region, expanding and completely obliterating the right maxillary sinus, and extending into the right nasal cavity, right sphenoid, ethmoid and frontal sinuses (Fig 4). The mass had caused complete occlusion of the right nasal cavity and was compressing the left nasal cavity, causing pressure atrophy of the bones. Further the mass was seen to extend into the right orbital region causing compression of the optic nerve resulting in unilateral exophthalmos. CT findings were suggestive of right maxillary region lymphoma causing secondary unilateral exophthalmos. Incisional biopsy from the right maxillary sinus approached through the right buccal mucosa i.r.t 15, 16 region revealed diffused B cell lymphoma of the maxillary sinus (Fig 5). Patient was referred for chemotherapy and radiotherapy.

DISCUSSION

Non-Hodgkin's lymphoma has a varied manner of presentation, response to therapy and prognosis. It rarely manifests as a primary malignancy in the head and neck region (> 1%) and may give an important clue for undiagnosed HIV infection, which accounts for 2% of oral neoplasms in patients with AIDS. It more commonly affects the middle aged and the elderly with a slight male preponderance. The gingival and palate regions are commonly affected. Secondary organ involvement along with the primary in the oral cavity is generally observed, but this was not observed in this patient.3

NHLs of the sinonasal tract are uncommon malignancies representing 3% to 5% of all malignancies, with NHL accounting for 60% of all lymphomas. B-cell lymphoma is predominant in paranasal sinuses. Tumour cells with positive T-cell markers (angiocentric lymphoma and peripheral T-cell lymphoma) are predominant in nasal cavities. In Western populations, lymphomas of the maxillary sinus are more common than in the nasal cavity.4 On the contrary, in Asian patients the nasal cavity is more common as a primary site than the maxillary sinus. More than 60% of NHLs of the head and neck occur in extra nodal sites, such as the paranasal sinuses, nasal cavity, oral cavity, salivary glands, and laryngopharynx.5 Early diagnosis of primary NHL of the nasal cavity and paranasal sinuses is difficult because this lesion develops in an anatomic space and expands toward the sinus, nasal cavity or nasopharynx, not usually causing symptoms in the early stages. Only after reaching a considerable size and involvement do the symptoms appear, and these may simulate other nasal or head and neck diseases.
The most common presenting symptoms of sinonasal lymphomas are nasal obstruction, epistaxis, headache, and unilateral facial, cheek, or nasal swelling. Other infrequent symptoms are diplopia or blurred vision, and nasal or cheek pain.2

Malignant lymphoma around the orbit is very rare, but it should be remembered that it can threaten vision by compressing the eyeball and/or optic nerve. Malignant lymphoma rarely arises in the Paranasal sinuses. Correct diagnosis of paranasal lymphoma is usually delayed. Because tumors at this site cause few early symptoms, the diagnosis is usually made at an advanced stage. Detailed investigation is often postponed at the initial stage and is delayed until the tumor causes clear symptoms. The abnormalities detected on X-ray films of the sinuses range from a soft tissue mass to orbital bone destruction. The most common sinus to be involved by malignancy is the maxillary sinus and squamous cell carcinoma is the most frequent malignant tumor.5

Indolent lymphomas are usually not curable and are treated with palliative therapy. Within 1 to 3 years, the disease usually progresses and requires therapy.6 Some studies have suggested that radiation alone provides good regional control at an early stage, while additional chemotherapy can be reserved for more extensive disease. However, other studies have shown that combined modality treatment with chemotherapy and loco-regional irradiation improves both disease-free and overall survival. The overall mortality rate is 55%. For effective treatment of these life-threatening malignant tumors, early detection by investigation of orbital symptoms may be important. In the present case patient developed unilateral loss of vision and exophthalmos. For correct diagnosis biopsy should be performed in patients with any unilateral intranasal or maxillary sinus lesion.5

CONCLUSION

Early diagnosis and staging are essential for effective treatment, and lymphomas must always be included in the differential diagnosis of lesions of the nasal cavity, paranasal and maxillary sinuses.

REFERENCES